A CASE

OF UNIVERSAL HYPEROSTOSIS, ASSOCIATED WITH OSTEOPOROSIS, WITH A DESCRIPTION OF THE SPECIMENS, by J. Ewing Mears, M. D., W. W. Keen, M. D., Harrison Allen, M. D., and William Pepper, M. D.

(*Read before the American Philosophical Society, Dec.* 2, 1870.)

The undersigned, to whom were referred the above specimens, presented by a friend to Dr. J. Ewing Mears, have carefully examined them, and have prepared the following Report:—

In the investigation of the subject, we have prepared as full a history of the case as could be obtained, a detailed account of the general anatomical characters of the disease, and of the peculiarities of each individual bone, as well as of the microscopic appearances, have consulted the works and periodicals in various languages accessible in this city, and have examined all the specimens contained in the Museums of the College of Physicians, Academy of Natural Sciences, University of Pennsylvania, Jefferson Medical College, and also the hospital and private collections in the city.

We have nowhere found specimens of this disease, or descriptions of such, at all equalling in extent and severity these here described.

The only similar case, though far less in degree and extent (skeleton imperfect), is found in Virchow's Archives, Vol. 43, 1868, p. 470, plate No. 12, although we have met with specimens and descriptions of skulls and bones which afford evidences of a limited development of the same disease.

Of the pathology of the disease, as well as of the anatomical appearances (116), we have found the best descriptions in Lobstein, Traité d'anatomie pathologique, Tom. II, p. 116; Boyer, Sur les Maladies, Chirurgicales, Tom. III, p. 571; Paget, Surg. Path. Eng. Ed., pp. 301–2, and fig. 40; Stanley on the Bones; S. Solly, Med.-Chir. Trans., Vol. 27; Förster Handbuch der Path. Anat., Bd. I, S. 249–52, and Bd. II, S. 850–4; R. Volkmann, in Pitha und Billroth's Handb. der Chirurgie, Bd. II, S. 249–58; Oeffinger, Virchow's Archiv, Bd. 43, S. 470; Haubner, Canstatt's Jahresbericht, 1854, Bd. 27, S. 23–4; Virchow, Die Krankhaften Geschwülste, Bd. II, Vorlesung XVII.

HISTORY OF THE CASE.

Fully recognizing the importance the history of the case has in the discussion of the Etiology and Pathology of the disease, we regret our inability to add any information to the statement given at the time of the presentation of the specimens, which is as follows:—

A. M. aet, 14, native of England—occupation farm boy—came from England to this country when very young—father died in November, 1862, of Phthisis, aged 57—mother died in 1867, cause of death not ascertained—has one brother and one sister, both young and healthy. In September, 1866, while engaged at work on the farm, noticed swelling

beginning in the face, and also, that in stooping, face felt puckered and wrinkled, while the effort to regain the erect position gave intense pain along the entire spinal column. Subsequently the fore-arms became sore and swollen, was placed under treatment, which was of such decided benefit that he thought himself entirely well, and in March, 1867, resumed his farm duties; about two months later the symptoms returned in an aggravated degree, the feet and then, in succession, the legs and thighs becoming enlarged and very painful; under the influence of constant bandaging the swelling diminished; his appetite became impaired, and he died from exhaustion Feb'y 8th, 1868. The treatment adopted was mostly of a tonic character.

DESCRIPTION OF THE SPECIMEN

(See Plates I. and II.)

I. WHAT BONES ARE WANTING.

We have the entire skeleton except the following bones:

Vertebræ—5th and 7th cervical.
1st and 10th dorsal.
2d lumbar.
2d and 4th sacral.
all the coceygeal.
Sternum, gladiolus and ensiform.
Hyoid bone.
Right Patella.
Hands, all wanting save the right scaphoid.
Feet, all wanting save left calcis.

Five metacarpal and metatarsal bones and eight phalanges are preserved, but, except the two metatarsals of the great toe, they can scarcely be designated, they are so greatly deformed.

II. THEIR CONDITION.

Unfortunately, by the prolonged boiling to which they were subjected before coming into our possession, the bones have lost probably all their animal matter, and are now almost as friable as if they had been burned. By removing the marrow, also, this has rendered the pathology of the disease much less clear and the microscopic examination much less valuable than it would otherwise have been. Moreover, it has removed probably all the gelatine, so that the chemical examination and the specific gravity would be worthless. Even the weights are, by reason of this misfortune, only of slight value. All the epiphyses too, except the coracoid process of the scapula, are separated from the shafts or bodies, and in some bones even integral parts are separated, *e. g.* the sacrum is divided into its component vertebræ and the innominate bone into two pieces. Many of the epiphyses are preserved, as will be indicated in describing each bone. The epiphyses have attached to them in many places the dried gelatinous articular cartilages of a transparent

brown color, and when an epiphysis has been incompletely ossified, the cartilaginous portion presents itself as a similar dried gelatinous mass. The ends of the shafts of the long bones are very ragged also, the cancellated substance being exposed and more or less broken.

III. WEIGHTS AND MEASUREMENTS.

In order to have some relative standard of weight, we have also weighed the bones of a girl of about seventeen. But, it must be observed, that all these but the scapula (which wanted the acromion epiphysis) were weighed with all the epiphyses. These healthy bones were rather slender, but were also longer than the diseased bones (the diseased and healthy femurs being 14½ and 16 inches respectively, excluding the lower epiphyses).

Diseased Bones.			*Healthy Bones.*		
Femur (without lower epiphysis)	8 oz.		Do (with all epiphyses)	8 oz.	3½ drms.
Tibia (" " ")	5½	"	Do " "	5 "	1½ "
Humerus (with all epiphyses)	3½	"	Do " "	2 "	3½ "
Radius (without epiphyses)	1½	"	Do " "		6 "
Ulna (without lower epiphysis)	2	"	Do " "		6½ "
Clavicle (" inner ")	½	"			
Scapula (" all epiphyses except coracoid)	1	"			
Fibula (without both epiphyses)	1½	"	Do " "		7½ "

The following are the measurements in circumference of the bones, the same healthy skeleton as before being used for comparison.

	Diseased Bones.	*Healthy Bones.*
Femur (middle),	5 in.	2½ in.
Tibia "	4⅜ "	2½ "
(at tubercle),	5⅝ "	4⅞ "
Humerus (middle),	4¼ "	2 "
(above condyles),	5 "	3¼ "
Radius (middle),	3½ "	1¼ "
(lower fourth),	4¼ "	1⅞ "
Ulna (" and middle),	3½ "	1⅛ "
(just below corocoid),	4⅛ "	1⅝ "
Fibula (lower fourth),	1¼ "	1⅜ "
Clavicle (acromial extremity),	3¼ "	2⅜ "

The following are the diameters. In general the original limits of the bone were pretty easily distinguished. The external line of demarcation in the femur and the posterior in the tibia are so indistinct that the diameters of the original bones are not wholly reliable. All the diameters are derived from longitudinal sections by a circular saw, and they are all at the middle unless otherwise stated.

	Original Bone.	*Diseased Bone.*	*Addition by Disease.*
Femur,	1 in.	$1\frac{1}{2}$ in.	$\frac{1}{2}$ in.
Tibia,	1 "	$1\frac{9}{16}$ "	$\frac{9}{16}$ "
Humerus,	$\frac{3}{4}$ "	$1\frac{3}{8}$ "	$\frac{5}{8}$ "
Radius (middle),	$\frac{7}{16}$ "	$1\frac{1}{16}$ "	$\frac{5}{8}$ "
(upper third),	$\frac{1}{2}$ "	$\frac{7}{8}$ "	$\frac{3}{8}$ "
(lower third),	$\frac{3}{4}$ "	$1\frac{1}{4}$ "	$\frac{1}{2}$ "
Ulna (middle),	$\frac{7}{16}$ "	1 "	$\frac{9}{16}$ "
(at coronoid),	$\frac{5}{8}$ "	$1\frac{1}{4}$ "	$\frac{5}{8}$ "
(lower third),	$\frac{3}{8}$ "	1 "	$\frac{5}{8}$ "
Metacarpal (great toe),	$\frac{1}{2}$ "	$\frac{11}{16}$ "	$\frac{3}{16}$ "
Phalanx "	$\frac{7}{16}$ "	$\frac{11}{16}$ "	$\frac{1}{4}$ "
Clavicle,	$\frac{3}{8}$ "	$\frac{3}{4}$ "	$\frac{3}{8}$ "
Fibula (upper & lower thirds)	$\frac{5}{16}$ "	$\frac{3}{4}$ "	$\frac{7}{16}$ "
Ilium (1 in. above acetabulum),	$\frac{9}{16}$ "	$\frac{7}{8}$ "	$\frac{5}{16}$ "

IV. General Description.

The bones which have suffered the most are the clavicle, humerus, radius, ulna, femur, tibia, fibula, metacarpals, or-tarsals and phalanges of both hand and foot. These are diseased in almost the entire length of their shafts. The radius and ulna have suffered rather more than any of the other bones just named. All the other bones of the trunk have suffered to some extent, those of the skull but very little or not at all.

Comparing the upper and the lower extremities, there is no appreciable difference in the violence of the disease.

Comparing the two sides externally, not only is there no difference in the extent and character of the disease, but there is the most remarkable symmetry of the corresponding, diseased bones, which may be traced even to details. (Figs. 9 and 10.) The disease begins and ends on both sides at corresponding points, it changes in character from simple porosity to the growth of osteophytes at corresponding points; if on one side the posterior part of the bone is most diseased, the same is true of the other side; if the osteophyte growth is continuous or interrupted on one bone (fibula Fig. 18) it is so on the opposite one; if one is unusually diseased at a tendinous or aponeurotic insertion, so is its mate; if a groove or a variation in color exist on the one side, the same will be found on the other side; even of single marked spiculæ of bone the same may be said; so that a description of one side will answer for both, minute differences being noted as they occur.

The main violence of the disease is expended on the shafts of the long bones. The epiphyses, of which the most important remain, *e. g.* those of the femur, tibia, humerus, &c., show we may almost say no disease. The lower epiphysis of the femur is slightly porous in the usually compact layer of the articular surface, but so fine is the porosity and so slight the disease that it would not be observed save on a most careful examination.

The other epiphyses show occasionally still slighter disease. Indeed it is a question whether this be not the result of the prolonged boiling. The bones of the trunk are but little affected except the sternum, which must have suffered severely, the manubrium being very porous and much thickened. The bones of the head are scarcely at all affected.

The point of greatest development of the disease varies with its character. 1°. The thickening is most developed in the middle of the shafts, and here generally the sclerosis is furthest advanced. (Figs. 3, 4, 5, 21.) 2°. The porosity is not noticeably greater in any particular parts of the shafts, but seems externally to be equally diffused. 3°. The osteophytes follow a marked law in their development. They are most developed where the muscles, aponeuroses, fasciae, &c., are attached, *e. g.* the linea aspera, interosseus ridges of the tibia and fibula, radius and ulna, the insertion of the deltoid, biceps and brachialis anticus, the condyloid ridges of the humerus. But it is not always true conversely, that where a large muscle is attached there must be a large osteophyte growth, *e. g.* there are none at the origins of the pectoralis major and sterno-mastoid, the supra- and infra-spinatus, the insertion of the quadratus femoris, &c. One class of exceptions is, however, to be noted, viz: that at the attachment of those muscles and ligaments that are connected to epiphyses, there is generally no disease, *e. g.* the muscular attachments to the greater and lesser trochanters, the greater and lesser tuberosities, tubercle of the tibia, the tuber ischii and nearly all the ligaments. The epiphyses and their attached parts are very nearly all quite free from disease, though it may be largely developed in their immediate neighborhood.

The direction of the nutritious artery seems to have had no influence on the development of the disease either in its extent or degree.

The porosity varies in its character, and usually any one bone will show all its varieties. 1°. The surface of the bone presents a very fine cribriform appearance, resembling pumice stone. When magnified six or eight times this is seen to consist of a stout network of bone perforated by numerous small foramina, which are generally tolerably circular, and do not communicate one with another. (Fig. 24.) 2°. It may be of a finer velvety appearance. This by the same power is seen to consist of the same network of bone, whose very large foramina or meshes now communicate and are therefore very irregular in form, while the ridges forming the bony net-work are very thin and form relatively high walls between the adjacent meshes. Sometimes these ridges assume a tolerably regular parallelism, giving a striated appearance to the part. 3°. A coarser appearance is often produced by a similar honey-combing with large foramina or meshes, deep and irregular, varying in size from a horse-hair to a line in diameter with the first or second variety existing in the intervening ridges. (See lower end of Humerus, Fig. 1.) 4°. The surface is often pierced more or less sparsely by small foramina about the size of a horse-hair. (Fig. 20.)

The osteophytes vary greatly also in their character. In shape they

are either pointed, flat, or clubbed, sessile or pedunculated. They frequently form larger or smaller scales, which cover more or less of the bone. They vary from the smallest size visible up to $\frac{3}{4}$ inch in length or $\frac{1}{2}$ inch square. They are often compound, smaller ones growing from larger ones as a base. Imbrication is not unusually a marked feature, and whether imbricated or not, their direction or "trend" almost always follows that of the fibres attached at that point. This is very marked where adjacent muscles run in different directions, *e. g.* the flexor attachments of the radius and ulna as contrasted with that of the pronater quadratus. (Figs. 11 and 15.) The grooves between the osteophytes have sometimes rolling edges, sometimes are as sharp as if cut with a knife, and often lie closely together and parallel; they are apparently made in many cases by the numerous small vessels. All of the osteophytes are more or less porous.

The color is usually normal, but in some places is of varying shades of brown.

On a section the whole bone is seen to be encased with a new formation of bone. This is true not only of the long bones but also of the scapulæ and ossa innominata. Viewing these bones and also many parts of the shafts of the long bones on the surface, one would suppose he had simply to do with the original thickened bone which had undergone this porous change. But a section shows that there is a complete new formation which is added layer after layer around the old bone. These layers (excepting where sclerosis has taken place) are separated by interspaces sometimes just appreciable to the eye, sometimes a quarter of an inch wide. (Fig. 17). The outer layer is often very thin, but presents to the eye that deceptive appearance of apparent compact tissue which has simply become porous. Where sclerosis has taken place or osteophytes are developed, of course the thickness of the outer layer is either greatly increased or else undeterminable. The other layers also vary in thickness from the development of the sclerosis from the thinnest possible to one or two lines. "These layers may sometimes be traced into continuity with those forming the healthy portion of the wall"* of the original bone, especially at the extremities of the shaft. (Fig. 4.) At these points, starting from the original compact tissue, the several layers of the encasing new formation gradually become more and more widely separated or new layers may appear, thus producing a very great thickening at the centre, while at the ends of the shaft the thickening gradually (sometimes suddenly) diminishes. The interspaces between the layers are sometimes for even an inch wholly void, but they are generally filled with intervening trabeculæ of bone, which form a cancellated tissue and also support the superimposed layer to which they are always *perpendicular*. To a very large extent these layers have been welded together by sclerosis, and sometimes the new growth and the original bone present no line of demarcation by which they can be distinguished. Where this solidification has taken place, the cut surface instead of the uniform ivory-like solidity of normal compact

* Paget. Surg. Path. Eng. Ed. 1863, p. 301–2, and fig. 40.

tissue presents a granular appearance, as if the cancelli of the interspaces were not solidly filled up. The process of sclerosis not infrequently dips down like a cone whose base is of considerable extent at the surface of the new growth and whose apex just touches, or is sometimes welded with the original compact tissue. (Fig. 21.) Eburnation has nowhere taken place.

The original bone, too, has undergone marked changes. Its limits are generally pretty well defined, but the compact tissue of which its wall once consisted, is now cancellated, to a greater or less degree (osteo-spongiosis). Sometimes all appearance of compact tissue, save a mere worm-eaten porous external film, has disappeared. Sometimes no cancellation appears, but the old and new growths are welded together. The cancelli of the once compact tissue of the old bone always run *parallel* with the *axis* of the bone, and are thus easily distinguished from those of the spaces between the laminæ of the new growth which run at *right angles* to the surface of the bone. (Figs. 4 and 17.) The old cancellated tissue has often very large cancelli and in some cases has disappeared, leaving a wider medullary canal than is normal.

The epiphyses do not appear materially altered on section.

V. Description of Individual Bones.

1. *Head.*—All the bones of the head are present, completely disarticulated. The spheno-occipital suture was not ossified. No sections were made of these bones, and the external appearances alone are described.

(a) Frontal. The roof of the orbit, especially in the fossæ for the lachrymal glands, is somewhat porous. Internally the porosity appears over various parts of the perpendicular portion. The irregular striated appearance from large numbers of fine grooves is marked, and sclerosis seems to have made considerable progress.

(b) Parietal. Externally slightly porous at the posterior border; internally also over, say one-fifth of the surface, corresponding to the protuberance.

(c) Occipital. Externally small scattered patches of porosity; internally the same change is limited to the superior fossæ and the groove for the left lateral sinus.

(d) Sphenoid. Porosity of external surface of greater wings, and also in most of the pterygoid plates, which are somewhat thickened.

(e) Temporal. Slightly porous and thickened externally on squamous portion, and in the glenoid cavity and in the grooves for both lateral sinuses.

(f) Sup. Max. Slightly porous on anterior surface, and at the tuberosity. The alveoli are reticulated so as to resemble almost the meshes of the pulmonary structure.

(g) Palate. Internally, slight porosity at the junction of the perpendicular and horizontal portions.

(h) Inf. Max. Ascending ramus markedly thickened, and porous internally and externally; most developed at the centre of the ramus;

body similarly affected, principally between the mental foramen and the external oblique line. Alveoli like those of the upper jaw. This bone has suffered more than any other bone of the skull.

Condition of Teeth.—The teeth were all present, and were carefully examined. They were very brittle, so as to break across with little difficulty (see *Micros. Ex.*), but presented no peculiarity of shape. The entire absence of the peculiar deformity of the incisors, noted by Hutchinson, of London, as characteristic of hereditary syphilis, is to be especially marked as it bears upon the question of causation of the morbid process.

2. *Vertebræ.*—The epiphysal plates of the bodies, and the epiphyses of the transverse and spinous processes, are all gone. In the dorsal region the groove between the three original parts in which the ossification takes place, is very deep, but they are all united more or less. This groove gradually disappears both above and below, none of the remaining cervical vertebræ showing it, while inferiorily it is visible as far as the first sacral. On section the body is not much thickened, and no line of demarcation exists. No sclerosis has taken place.

(a) Cervical. Scarcely noticeable porosity of the anterior surface of body. Posterior arch of atlas is unusually thick and dense.

(b) Dorsal. Marked porosity of external surface of body, which is elevated above the surface left by the removal of the slightly overlapping epiphysal plates, about one-half a line to a line. Spinous processes slightly porous.

(c) Lumbar and Sacral. Same as dorsal; the porosity of spinous processes being more marked.

3. *Sternum and Ribs.*—(a) The manubrium only is present, and is very thick and porous. No osteophytes.

(b) The ribs have lost all their epiphyses. They are not affected on the external surface, save slightly in one or two instances. On the pleural surface they are all porous, and often a little thickened. For about one inch from the head the entire bone is thickened and porous.

4. *Upper Extremities.*—(a) Clavicle. The sternal epiphysis is wanting. Where the surface for the articulation with the acromion should be, there is on each side an oval cup-like depression $\frac{1}{2} \times \frac{1}{4}$ in. and $\frac{1}{8}$ in. deep. (Fig. 13.) Its walls are perpendicular, its floor flat, and both are covered with a thin layer of compact tissue resembling that which covers all the ends of the diaphyses of the other bones next the epiphysal cartilage. It was filled, when first seen, with a small mass of dried tissue resembling the epiphysal cartilage already described. Possibly it may have been an unusual third centre of ossification for this bone. It was occupied, certainly, by some substance separate from the shaft of the clavicle, either a third centre of ossification, or a projecting piece of the acromion. If the former, it is a very unusual place for a supernumerary epiphysis.

The whole bone is thickened to about twice its normal width, and its surface is coarsely porous throughout. At the insertion of the ligaments on the under surface, the porosity is quite fine and velvety. At the inner half of the origin of the deltoid there are twelve to twenty stout and well

developed osteophytes. A few also exist at the middle of the insertion of the trapezius. The section shows the original bone distinct from the new growth at all points. The laminæ of the new growth are very distinct at most points. A large part of those of the under surface are more or less closely united by partial sclerosis. The original bony tissue is relatively but little altered.

(b) Scapula. The coracoid process is one-third united to the bone, but wants the epiphysis developed on it at about seventeen years of age. All the other epiphyses are absent.

The bone is porous throughout, save at the centre of the infra-spinous fossa; generally of the coarse variety, but very fine in certain spots. The whole bone is somewhat thickened, as can be seen without any section, at its posterior border and on the spine. (Fig. 23.) The latter being to a great extent denuded of the outermost compact, yet porous layer of the new growth, shows the reticulated trabeculæ which supported it, and through their meshes the old external compact layer of the original bone now all worm eaten and very thin. This is especially well seen at the two extremities of the spine. The axillary border of the bone is three or four times as thick as is normal, has a few coarse osteophytes, and a very deep and wide groove for the dorsalis scapulæ artery.

(c) Humerus. All the epiphyses are preserved except that of the internal condyle. The trochlear surface projects only to a level with the radial.

The whole shaft (Figs. 1 and 2) is involved in the disease, the least at the upper fourth, the other three-fourths being about alike. The porosity is almost wholly very fine or velvety. About two inches below the head, at the insertion of the Pect. maj., the anterior bicipital ridge is greatly thickened (especially on the right side). It is continuous with a very large elevated surface ($2 \times 1\frac{1}{2}$ in.) at the insertion of the deltoid. This is covered with a large mass of not very large porous osteophytes whose trend is generally upwards. One (r. side) or two (left) large flat imbricated osteophytes mark the posterior lip of the bicipital groove. At the musculo-spiral groove, which is well marked, the bone is finely porous, but presents no osteophytes. At the lower third, anteriorly, the bone presents numerous osteophytes, sometimes single, but generally in groups. They are sessile, porous, and in some cases imbricated; their trend is generally downwards, except just above the epiphysis, where they are at right angles to the bone. The two condyloid ridges, especially the inner, are greatly diseased. The external ridge (especially on the right side) has several large porous sessile outgrowths with intervening grooves, the largest groove about corresponding in position to the anastomotica magna artery. The internal ridge up to the insertion of the coraco-brachialis is covered with large knobby and porous, imbricated osteophytes, continuous with a similar remarkable growth on the posterior surface of the bone, covering the origin of the internal head of the triceps, which extends to the musculo-spiral groove above, and fades into simple porosity externally. At the origin of the external head of the triceps, there is also a

marked elevation covered with pointed osteophytes, and continuous with that of the deltoid insertion. The trend of all these osteophytes is downwards, and their color (especially on the right side) is a light brown.

The section (Figs. 3 and 4) shows the outline of the old bone obliterated in the lower third, and only faintly visible in the upper two-thirds anteriorly. In the posterior upper two-thirds the laminæ of the new growth are admirably shown, though even here the sclerosis is in some parts far advanced. The original compact wall in the superior one-third, anteriorily, and two-thirds posteriorly, has almost disappeared, the cancellation (spongiosis) is so great, and it is a typical illustration of this process in various stages. The original cancellated structure is either fragmentary, its cancelli being very large, or else it has entirely vanished, leaving an enlarged medullary canal.

(d) Radius (Figs. 9, 10 and 11.) All the epiphyses are gone, save the left upper one. Instead of being rounded externally, and showing a sharp interosseous ridge internally, it is almost cylindrical, increasing in diameter from above downwards. At the bicipital tubercle there is a crest of curved osteophytes under which, as in a cave, the tendon of the biceps was inserted. The oblique line is marked by a series of knobby, porous, slightly imbricated osteophytes, whose trend is downwards and inwards till they reach the insertion of the pronator teres, where their size increases, and their trend is upwards and outwards. The interosseous border is rounded off and marked by a series of deeply imbricated laminated osteophytes, all trending downwards, resembling a rounded surface deeply grooved by oblique parallel cuts of a thin saw. Where the pronater quadratus was attached, a large number of osteophytes exist in ridges, which run latterally. All the rest of the bone is thickened and porous, and where the muscles took origin, is covered with porous osteophytes.

In section (Fig. 8) the outlines of the original bone are visible throughout; the laminæ of the new growth are marked; the sclerosis is in various stages, and anteriorly for some two inches the new and old growths are almost welded together. The old compact tissue is wholly changed to spongy, and the medullary canal is increased in size.

(e) Ulna (Figs. 14 and 15.) The lower epiyhyses are absent. Like the radius, the ulna is involved in its whole length, and is about twice its normal diameter. At the insertion of the brachialis anticus, a cup-like depression surrounded by an elevated ridge of osteophytes, exists, somewhat similar to that on the bicipital tubercle of the radius. The anterior surface is covered with small porous osteophytes, with a slight downward imbrication. At the attachment of the pronater quadratus they become more marked in their development, and the imbrication is external. The interossecus ridge is rounded off and marked, as in the radius, but with several unusually large and deeply imbricated osteophytes with a deep groove, probably that of the interosseous artery. Externally a brown discoloration is seen, which is the most noticeable on the right side. Posteriorly the bone is coarsely

porous, but very few osteophytes exist, save on the lower third. The intervening grooves run transversely, but are neither deeply nor sharply cut.

On section (Figs. 16 and 17) the line of the original bone can be distinguished throughout; the laminæ of the new growth are very marked; the sclorosis has welded together all the new layers anteriorly, and at the junction of the upper and middle thirds, the new and old growths are almost melted together both anteriorly and posteriorly. The interspace between the old bone and the first new lamina reaches one-fourth of an inch in width just below the olecranum, and the distinction between the perpendicular trabeculæ filling it up, and the longitudinal cancelli of the once compact tissue of the old bone is very marked. The medullary canal is scarcely, if at all, enlarged, and, indeed, at the point of greatest sclerosis above named, the same process seems to have invaded the canal itself.

5. *Lower Extremities.*—(a) Innominate Bones. The ilium is separated from the ischium and pubes, which are firmly and indistinguishably united together at their rami, but at the acetabulum are distinct. The Y-shaped piece uniting them is preserved, and is loose on both sides. All the other epiphyses are missing. The bones are porous throughout but not to a marked degree. The thickening varies from $\frac{1}{8}$ to $\frac{3}{8}$ of an inch, being greatest just above the acetabulum. On the ischium and pubes no osteophytes exist, save one small lamina on the body of the right pubes. The ilium is free from them except above the acetabulum for a considerable space, on and around the reflected origin of the rectus, where large and strong osteophytes exist, with a trend inwards and upwards.

On section of the ilium, (Fig. 12) the external surfaces, which otherwise would be thought to be the porous surface of the original bone, are seen to be the outer layer of the new growth. The original compact tissue has undergone spongiosis to a great extent. Sclerosis is furthest advanced just above the acetabulum.

(b) Femur (Figs. 6 and 7). All the epiphyses are separated. Both heads and great trochanters and the left lower epiphysis are preserved. The latter shows some very slight porosity, as already noticed.

Anteriorly the inter-trochanteric line is marked by a well developed growth of short, thick, rather acuminate osteophytes, separated by grooves running in the axis of the neck. A similar line of more slender imbricated osteophytes runs parallel to the base of the great trochanter and trends toward it. These two lines form the letter A. Immediately within this letter A (especially on the left side) the trend of all the osteophytes turns sharply downwards and so continues to the lower $\frac{1}{4}$ of the bone, where they are perpendicular. They are not very marked in their development. Just above the end of the shaft, however, they form an overlapping sheath to the bone. In the middle of the right femur an aperture ($1\frac{1}{4} \times \frac{3}{8}$ inches) exists in the ensheating new growth, disclosing

to view the original but altered bone. Posteriorly the osteophyte growth extends from half an inch below the lesser trochanter to within $2\frac{1}{2}$ inches of the end of the shaft, and the same sheath-like appearance is very noticeable at its two extremities. Where not covered with osteophytes, the shaft is very finely porous and thickened. All the central two-thirds of the shaft is one vast mass of large, irregular, porous osteophytes. Their direction is not constant, but is in general downwards, and their shapes are very varied. The mass extends for about eight inches, along what was the tolerably sharp linea aspera, but is now about $\frac{3}{4}$ in. wide and about $\frac{3}{8}$ inch thick. The posterior inter-trochanteric line and great trochanter are not affected, except a slight porosity in the former. The lesser trochanter is wanting, but for $\frac{1}{2}$ in. around it there is no disease beyond some porosity save one squamous osteophyte on the right side.

On section (Fig. 5) in the axis of the head and great trochanter the outline of the old bone is not to be made out save internally, and then only imperfectly. The old and new growths are almost everywhere indistinguishably welded together. The laminæ of the new growth, too, are welded together save at a very few points. The old compact wall is still solid, but it looks granular and does not present the ivory-like solidity of normal compact tissue. The medullary canal is somewhat enlarged at the expense of its walls. At the lower extremity the trabeculæ of the cancellated substance are normal, but in the head and neck the arches for mechanical support are much less distinctly marked than is usual.

(c) Patella. The right patella is missing. The anterior surface of the left shows a few osteophytes trending downwards.

(d) Tibia. Both the upper epiphyses are preserved. The whole bone is greatly diseased and thickened to about twice its usual diameter. The tubercle is slightly thickened and presents a ragged edge above for articulation with the epiphyses, but the greater part of the tubercle being developed from the epiphyses the disease is not very marked. The crest is rounded in its whole length and porous. The internal sub-cutaneous surface presents marked swelling and porosity. There is but little osteophyte growth, and it is generally in the laminæ except at the sartorius insertion, where it is more developed. A number of deep grooves exist, generally longitudinal in their direction and most marked at the upper third. The posterior and external surfaces are covered with a warty growth of porous osteophytes which attain their greatest development at the interosseous border and especially at the oblique line. The general trend of all this growth is downwards. Grooves for the vessels are frequent and tolerably deep.

On section (antero-posterior) the outline of the old bone is distinct at the extremities, but in the central two-thirds it is barely visible in front and wholly lost behind, the sclerosis at this part having welded together all the laminæ of the new growth and the original bone. Even at the extremities the new laminæ are not very marked. The rarefaction of the original compact substance is of course therefore not marked. The medullary canal if at all altered is narrowed by the encroaching sclerosis.

(e) Fibula (Fig. 18). Its shaft alone is preserved, and its axis is slightly bent inwards. The whole bone is encased in a newly formed osseous growth which is sometimes simply porous, and is covered with sometimes an interrupted, sometimes a continuous, growth of warty osteophytes, all more or less porous. Posteriorly and internally this growth is most developed, the trend being downwards. The lower subcutaneous surface is greatly thickened and finely porous, but has no osteophytes.

On section the bone is doubled in its diameter, the outline of the original bone being only visible in about one-half of its extent, the sclerosis obscuring it at other points. The original compact tissue is rarefied by spongiosis, and the medullary canal is somewhat widened.

(f) Hand and Foot. They are considered together, as some of the bones are indistinguishable, and moreover, in general the same description applies to both.

(1.) Right scaphoid of hand. Not diseased.

(2.) Left calcis. Porous and enlarged throughout. Porous osteophytes are seen at the attachments of the tendinous sheaths internally, and one large flat one on the inferior surface. The epiphysis for the attachment of the tendo achillis is preserved, but shows no disease.

(3.) Metacarpus, metatarsus and phalanges. Two of the phalanges have their epiphyses attached but not united by ossification. The epiphyses are not diseased. No other epiphyses are preserved. All these bones suffer by far to the greatest extent in the centre, not at all at the head (viewed externally), and but little at the base, and the new growth is five or six times as thick on the dorsum as on the opposite surface. No osteophytes exist save on one of the metatarsal bones and at the ridges for the flexor sheaths of three of the phalanges.

On section (Fig. 21), compare also Fig. 22, the outlines of the old bones are very readily seen, the apex of the conical sclerosis having, at points, just touched the surface of the old bone. The original wall of compact tissue is wholly rarefied by spongiosis, and the cancelli of the new and old bone are readily distinguished by the different directions of their axes. The normal compact wall of the phalanges being very thick relatively, the changes in it are the more marked. Sclerosis has invaded from half to two-thirds of the new growth. The head of the bone is also markedly rarefied by spongiosis.

MICROSCOPIC EXAMINATION.

The specimens from which the following description and wood-cuts were made, were prepared with his well known skill by Dr. J. H. McQuillen. They consisted of a transverse section through the thickened wall of a phalanx, embracing the thickness of the layers superimposed by the periosteum, but not of the entire original compact layer of the bone, and of a transverse section through the right canine tooth.

The section of the phalanx (Fig. A) exhibited a quite compact osseous structure—the Haversian canals being for the most part round, and rather small, though in some places they were irregularly shaped or oval, and larger. The intervening bone lamellæ were of unusual thickness,

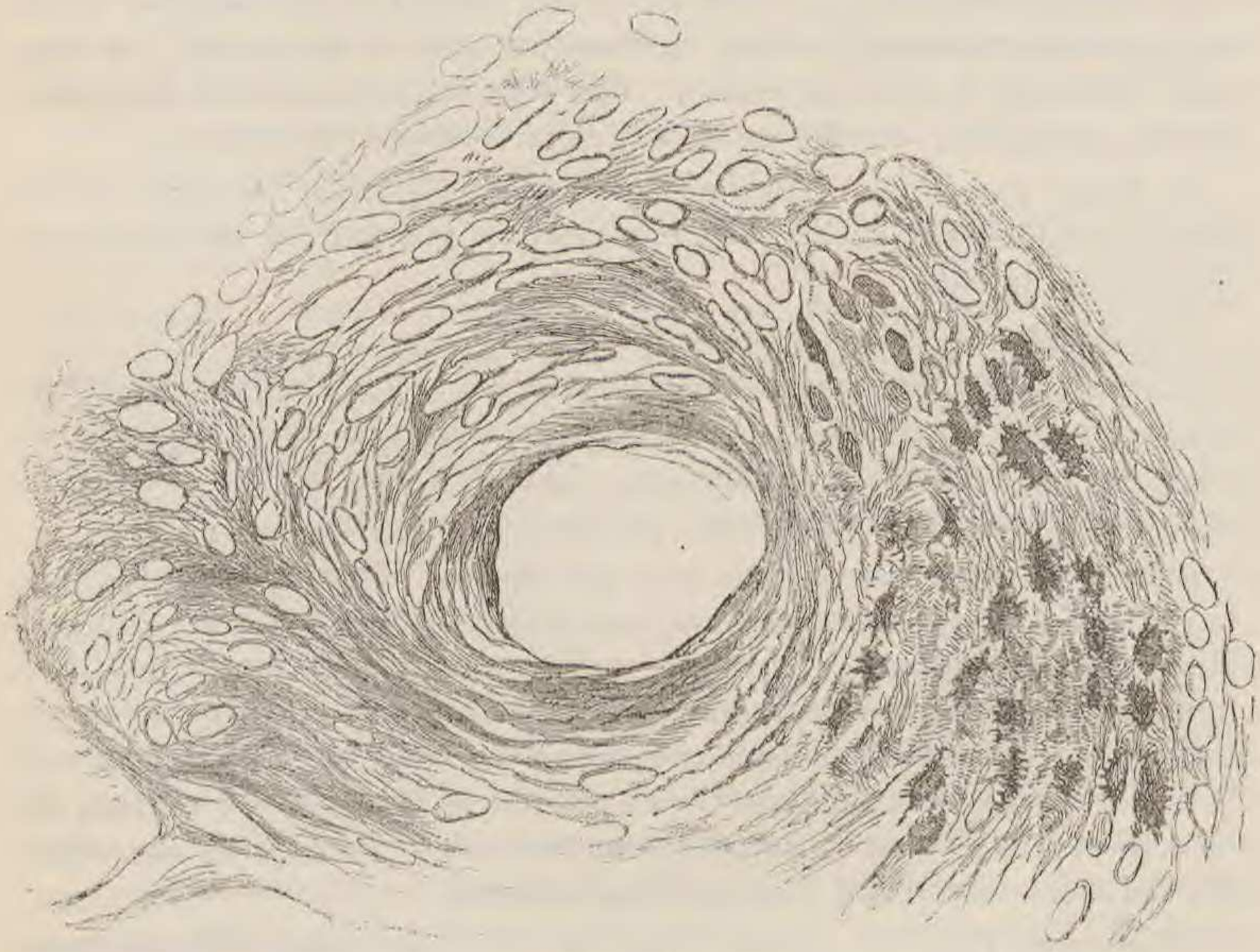

Fig. A.

and presented in the majority of cases, bone corpuscles with canaliculi. In some cases, however, no bone corpuscles were present, and the lamellæ appeared to be merely calcified by saturation with bone salts.

The bone corpuscles were small and often indistinct; in some places they were unusually round, but in others they presented the normal elongated shape; their canaliculi were invariably very poorly developed, and often could not be discovered.

With regard to the mode of arrangement of the bone lamellæ, they were always developed concentrically with (parallel to the walls of) the Haversian canals, and in no instance were any lamellæ found whose direction was parallel to the external surface of the shaft.

The section of the tooth (Fig. B) showed the existence of numerous irregularly shaped, branching lacunæ in the dentine near the marginal layer of enamel. These spaces were of various sizes and intercepted the

FIG. B.

course of a varying number of dental tubuli. They indicated unquestionably either an arrest of the process of calcification of the dentine, or of the resorption of calcareous matter already deposited, conditions which are also present in the true bony tissue. They are identical with the so-called *interglobular spaces* first described by Kölliker and carefully studied by Dr. McQuillen of this city, who has published (Dental Cosmos, N. S. Vol. VIII, No. 3, pg. 113,) several excellent illustrations of them.

PATHOLOGY OF THE DISEASE.

Having thus described the gross and minute features of these bones, we would hazard the following remarks in regard to the nature and cause of the pathological process :—

In the first place it is to be observed that three separate processes, or at least three distinct stages of the same process, are represented in different parts of the skeleton, or even, in some instances, in single bones. These stages are :

First. Internal Osteoporosis of the original osseous tissue. 2d. External Hyperostosis, due to successive attacks of Periostitis, both of which processes are present in varying proportion in almost all the bones, and 3d. Secondary Induration.

1. It is especially in regard to the explanation of the internal osteoporosis, which constitutes so marked a feature of these specimens, that we regret the absence of any careful examination of the bones in their recent condition. In their present state, it is only possible to describe the degree to which this rarefaction of the osseous tissue has occurred, but it is evident that such changes might be produced by very varied alterations of the medulla and bone corpuscles. Thus, among the recognized causes of osteoporosis, may be mentioned syphilis, scrofula, rheumatism; and, in addition, we must add that both osteomalacia and simple ostitis produce changes in the bones which, after the specimen has been boiled, and the organic matter entirely removed, are not to be distingushed from the effects of the first mentioned diseases. In each case, under the action of the morbid irritant, whether purely external and local, or internal and constitutional, there is more or less rapid proliferation of the essential vital elements of the osseous tissue, called bone corpuscles or cells. At the same time, the bony laminæ surrounding the Haversian canals, and the walls of the lacunæ, are progressively deprived of their calcareous salts and removed, while the enlarged spaces thus produced are filled by the constantly growing cellular elements. The manner in which this removal of the calcareous salts is effected has been, and indeed remains, a subject of much discussion. The first idea which seems to have been entertained may be inferred from the name, eccentric atrophy, which was given to many specimens of osteoporosis, under the belief that the bony lamellæ were thinned and pushed asunder by the centrifugal pressure of the growing medulla. There is, however, no evidence whatever in favor of such a mechanical explanation, and this hypothesis has justly been almost universally abandoned.

By far the most plausible explanation which has been advanced appears to be that the removal of the calcareous salts, the first essential step in the destruction of the bony lamellæ, is due to the solvent action of some acid elaborated by the bone cells during the inflammatory process. According to Weber, their removal is not due to the direct action of any acid, but is owing to a gradual conversion of the insoluble *tri-basic* phosphate of lime into the more soluble *bi-basic* salt.

As this feature of rarefaction is, however, common to so many diseases of the bones, it is evident that the most characteristic results of such diseases are to be rather found in the condition of the bone cells, and in the characters of the morbid product which has resulted from their multiplication. And it is to be trusted, that by careful chemical and microscopical study of these, such peculiarities will be discovered as will enable us to distinguish with certainty in recent bones the various morbid changes.

Heretofore the majority of observers have limited themselves either to a description of the dried bone, after maceration or boiling, or at most, of the general characters of the medulla with which its cancelli are filled. And it results from this superficial mode of study, that there is as yet but

little exact knowledge of the really essential changes which the organic, active portions of bones undergo in disease.

Virchow, who was among the first to examine microscopically the condition of the bone cells in ostitis and some other diseases attended with rarefaction of the bone tissue, (Uber parenchymatöse Entzündung; Virchow's Archiv. Bd. IV. Hft. 2: 1852, b. s. 301 to 311,) formerly regarded the process as essentially a degenerative one, due to the fatty degeneration of the bone corpuscles and the subsequent softening and removal of the area depending on these cells. We have already, however, stated the view which appears most plausible in regard to the removal of the calcareous salts, and so far from fatty degeneration of the bone corpuscles being a constant feature in the different forms of osteoporosis, it would appear from the careful researches of Ranvier (Archives d' Anat. et Phys., Norm. et Path., No. 1, 1868, page 69), that this condition of the cells is altogether characteristic of caries and limited to that morbid process. On the other hand, there is every reason to presume that these cells are influenced by various morbid causes, (inflammation, syphilis, rheumatism, gout, scrofula, &c.) in the same way as the other tissues of the body, and give rise to products more or less characteristic of the diseased action present.

The history of the present case would appear to indicate that the nature of the disease was a rheumatic or scrofulous inflammation, but beyond this mere supposition we are prevented from advancing by the absence of any chemical and microscopical examination of the recent bones.

We would here again call attention to the marked peculiarity of the porotic bones, fully described at pages 24 and 25, and figs. 4 and 17, although we are unable to suggest any plausible explanation of the invariably parallel arrangement of the meshes of the porotic bone, and of the equally uniform vertical arrangement of the meshes of the new-formed sub-periosteal layers.

2. Another important appearance present in the bones here described, and indeed one which is as marked and wide-spread as the osteoporosis, is the extensive development of bone upon the exterior of the original shafts. In our description of the skeleton, we have already noted the peculiarities of these sub-periosteal growths, and it will be remembered that they are in every instance limited to the body or shaft of the bones, and never extend on to the epiphyses, and that they usually present several thin laminæ of imperfectly compact bone, parallel to the shaft and separated from it and from each other by more or less wide interspaces usually occupied by coarse cancellated tissue.

It is undoubtedly from the examination of such specimens as this that the mistaken idea arose that the lamellæ, of which the original compact shaft was formed, had been pushed asunder by the great enlargement of its cancelli. It will, however, be seen from our description that the ap-

pearances contradict any such supposition, and clearly show that while in the cancellated and imperfectly compact tissue of the original shaft a process of rarefaction (osteoporosis) has been advancing by atrophy of the bony lamellæ, there has also been an active process of periostitis resulting in the formation of thick layers of new bone on the exterior.

Another means of distinguishing the line of demarcation between the original shafts and the new-formed layers, is the abrupt change in the direction of the cancelli already referred to.

It is evident, also, that the periostitis has not been uniformly continuous, but that for a variable time its intensity was such that the inflammatory product was capable of but imperfect ossification, and remained as cancellous tissue; while at irregular intervals thin layers of imperfectly compact tissue have been formed. The occurrence of this long standing, but not uniform process of periostitis ossificans appears to account, in every instance, for the changes observed on the exterior of the original shafts. In addition to this uniform hyperostosis, it will be observed from the description (see pp. 23, 24) that the same process of periostitis has given rise to varied forms of porous osteophytes.

3. In some places, however, it is evident that a still further change has occurred, consisting in the gradual conversion of the cancellous tissue into compact bone. This process of consecutive or secondary induration is most marked in the layers of bone formed by the periosteum; though it is present in the shafts of the tibiæ, femora and some other bones. It is manifestly impossible to determine accurately the portions which have been rendered compact by this process, but the disposition of the successive layers of new-formed bone is, in general, so much like that above described, that we are inclined to regard all the areas of compact bone of any considerable thickness as due to this secoudary change.

It would, indeed, appear but probable that as the high degree of inflammation, under which the layer of cancellous tissue had been formed, subsided, there should be a tendency to the formation of successive layers of bone on the interior of the walls of the cancelli. It is especially in connection with this point that the result of the microscopical examination of the sub-periosteal layers is of so much interest. It will be observed (see Fig. A.) that in the newly-formed compact bony tissue, the lamellæ are arranged concentrically around the vascular canals; a mode of arrangement which strongly points to the occurrence of the process of consecutive induration, as we have above described.

The specimens are deposited in the Museum of the College of Physicians.

J. EWING MEARS, M. D.
WM. W. KEEN, M. D.
HARRISON ALLEN, M. D.
WM. PEPPER, M. D.